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## Life with Cortical Dysplasia. A case study of a sixth grader.

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# Life with Cortical Dysplasia. A case study of a sixth grader.

## **Abstract**

Cortical Dysplasia is a rare brain malformation that results in multiple life challenges such as hemiparesis (cerebral palsy on half of the body), and seizures. For some people, with medication and life long therapies, the side effects dealing with cortical dysplasia can be minimized. This can lead to those living a rather normal life. In some cases, more drastic measures have to be taken. A hemispherectomy is the removal of the entire hemisphere of the brain, typically the malformed side. The idea of this surgery is to remove the part of the brain causing life-threatening problems. In cases for those suffering from cortical dysplasia, the idea is to remove the side of the brain causing seizures. In order for one to qualify for this surgery, a number of tests need to be implemented. The most important test deals with the strength of the seizures without any medication. If the strength of the seizure passes that test, then that person will then qualify for the surgery. Connor Smith (pseudonym) suffers from cortical dysplasia. Due to the strength and damage of the seizures cortical dysplasia, Connor was able to undergo the hemispherectomy surgery. The challenges that Connor faces daily cause him to be an inspirational story to remind others that special education does not just mean one learns differently, but that their care and academics need to be individualized in order to create success.

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Cortical Dysplasia is a rare brain malformation that results in multiple life challenges such as hemiparesis (cerebral palsy on half of the body), and seizures. For some people, with medication and life long therapies, the side effects dealing with cortical dysplasia can be minimized. This can lead to those living a rather normal life. In some cases, more drastic measures have to be taken.

A hemispherectomy is the removal of the entire hemisphere of the brain, typically the malformed side. The idea of this surgery is to remove the part of the brain causing life-threatening problems. In cases for those suffering from cortical dysplasia, the idea is to remove the side of the brain causing seizures. In order for one to qualify for this surgery, a number of tests need to be implemented. The most important test deals with the strength of the seizures without any medication. If the strength of the seizure passes that test, then that person will then qualify for the surgery.

Connor Smith (pseudonym) suffers from cortical dysplasia. Due to the strength and damage of the seizures cortical dysplasia, Connor was able to undergo the hemispherectomy surgery. The challenges that Connor faces daily cause him to be an inspirational story to remind others that special education does not just mean one learns differently, but that their care and academics need to be individualized in order to create success.

## Literature Review

Epilepsy is a major neurological disorder, the symptoms of which are preventable and controllable to some extent (Bhalla, Godet, Druet-Cabanac, Preux 2011). Multiple causes can create different forms of epilepsy. Those causes are either predominately developmental/genetic or predominantly acquired throughout time. Some of those causes include brain tumors, traumatic brain injury, bacterial infections in the brain, neurocysticercosis, cerebral malaria, stroke, cerebral malformations, Alzheimer's disease, multiple sclerosis and much more (Bhalla 2011). All can cause severe life changing cases of epilepsy that can only be assisted with medications or even surgery such as hemispherectomy and a multilobar. Fortunately, many patients that receive the corrective surgery have high success rates of a seizure free lifestyle. Those that are not as fortunate have to come up with alternative ways to create a positive lifestyle for themselves, while still dealing with the affects of epilepsy.

There are many people with epilepsy that are classified as having traumatic brain injury, with incidents of seizures increased seventeen-fold for severe TBI and by a factor of 1.5 for even mild TBI without skull fracture or prolonged loss of consciousness (Matsumoto, Caplan, McArthur, Forgey, Yudovin & Giza, 2013). According to some research, TBI affects more than 475,000 children under 15 years of age annually, with the highest rates observed in children 0-4 and a second peak in adolescence (Matsumoto 2013). Research has shown that disorders called focal cortical dysplasia (FCD) and mild malformation of cortical development (mMCD) are common causes of epilepsy, cognitive disability, and other neurological disorders as well (Krsek, Johodova, Maton, Jayakar, Dean, Korman, Rey, Dunoyer, Vinters, Resnick & Duchowny 2010).

Cortical dysplasia can be defined as a spectrum of derangements in development of the neocortex associated with a range of morphologic features (Chung, Lee & Kim 2005). In simpler terms, according to Cincinnati Children's Hospital, it means the top layer of the brain did not form properly, and as the brain naturally develops the neurons spread to the outward cerebral cortex. When the neurons fall or get misplaced, it causes signals to "misfire," which can cause a seizure (*Cortical Dysplasia in Children*). According to Cincinnati Children's Hospital, there are three types of FCD. Type I is difficult to see in a brain scan, and often found in patients that do not start having seizures until they are adults. This usually involves the temporal lobe of the brain. Type II is a more severe form of FCD, and is more often found in children. It typically involves both the temporal and frontal lobe of the brain. Type III involves damage in another part of the brain, which could possibly be due to a brain injury early in life (*Cortical Dysplasia in Children*). Each type creates different forms of epilepsy per person some can be mild and some are severe.

As revealed by magnetic resonance imaging (MRI), cortical dysplasia is the second most common cause of medically refractory chronic partial epilepsy (Chung 2005). Those diagnosed with cortical dysplasia suffer from seizures that can be monthly, weekly, or even daily, depending on the severity. It can also be the consequence of a wide range of disorders affecting the brain including tumors and various non-epileptic lesions (Miyata, Hori & Vinters 2013), all of which play a huge role in how people diagnosed with this disorder live their lives. People dealing with cortical dysplasia have struggles they deal with on a daily basis, from academic, to physical or emotional issues.

Patients suffering from cortical dysplasia are told that total resection of the lesion has been reported to be the predictor for a good seizure outcome (Daniel, Meagher-

Villemure, Roulet & Villemure 2004). About 70 patients with cortical dysplasia have focal or lobar resection, and these cases are often older patients, and the remainder either have a multilobar operation, which is the removal of parts of two or more lobes in the brain, or hemisphere operation, which is the removal of the entire half of the brain, at an earlier age.

The type of hemispherectomy (right or left) can determine the type of outcome for patients. Studies in adults have shown that left hemispherectomy leads to differential impairment in language while the right hemispherectomy, to particular loss of spatial skills, in children; however, this pattern has not been generally reported, at least not in a straightforward way (Pulsifer, Brandt, Salorio, Vining, Carson, & Freeman 2004). Reports indicate that on average 62% of patients with cortical dysplasia are reported seizure-free after surgery (Hauptman & Mathern 2012).

Focal cortical dysplasia (FCD) was first described in a seminal 1971 paper reporting the neuropathologic features of ten patients with drug resistant focal epilepsy that had undergone epilepsy surgery. The altered cortex was characterized by the presence of an excess of large, randomly oriented, aberrant pyramidal neurons, often crowded together. Most cases were also characterized by the presence of numerous large, malformed cells of uncertain origin that were frequently multinucleated, and had an excess of opalescent and pseudopodic cytoplasm. Although some of these morphologic features resembled those of tuberous sclerosis, the authors interpreted their data as identifying a distinct type of cortical malformations associated with severe seizures (Finardi, Colchiahi, Castana, Locatelli, Nobili, Fratelli, Bramerio, LoRusso, & Battaglia 2013).

FCD is now extensively used to refer to focal abnormalities of cortical structure of various types. The international League Against Epilepsy just proposed a new FCD

classification that considers electroclinical imaging and neuropathologic features.

Neuropathologic analysis type II FCD is characterized by disrupted cortical lamination and presence of large dysmorphic neurons and balloon cells. FCD is also characterized by the distinctive clinical features, such as early age epilepsy onset, high seizure frequency, episodes of status epileptics and continuous spiking stereo-EEG (stereo-electroencephalogram) during testing (Finardi 2013). Electroencephalogram is a test that detects electrical brain activity using small metal discs attached to the scalp according to the Mayo Clinic (*EEG Electroencephalogram*).

The efforts underlying this severe form of epilepsy are not completely understood. It is likely that increased glutamatergic inputs or altered expression or function of the N-methyl D-aspartate (NMDA) (predominate molecular device for controlling synaptic plasticity and memory function) receptor are involved, since NR2A/B subunit levels are increased in surgical specimens from patients with FCD, and epileptogenic activity on cortical slices from such patients is sensitive to specific inhibitors of NR2B.

While it is generally accepted that acute seizures could damage the brain, there is little evidence that epilepsy can progressively alter brain structure and function. However, the evidence has recently reported that epilepsy can alter brain structure and function due to findings from a rat model of epileptogenic FCD characterized by altered NMDA expression and progressive structural abnormalities. These findings suggest that status epileptic and subsequent seizures actuate a pathologic process that modifies the structure and function of the malformed brain. All of these findings lead to researchers completing studies on patients dealing with FCD (Finardi 2013).



In a present study, researchers examined whether cortical changes associated with epilepsy were also present in human epileptogenic cortical malformations. A case study completed on eight patients with type II FCD and variable duration of epilepsy compared morphologic features between cortical areas at the origin of epileptic discharges with those in adjacent areas and non-FCD controls (Finardi 2013). Eight patients with type II FCD, two epileptic patients without cortical dysplasia and seven non-epileptic controls with non-invasive brain tumors of comparable age were examined throughout this case study. The patients were surgically treated at the C. Munari Epilepsy Surgery Center, Niguarda General Hospital, or the Neurosurgery Department of the C. Besta Neurological Institute. For epileptic patients, high-resolution MRI and electroclinical investigations including stereo-EEG (in five cases) were used to identify the epileptogenic areas to be ablated. In all cases, cerebral specimens were removed for strictly therapeutic reasons, after informed consent had been obtained for the ablation and used of removed material for research (Finardi 2013).

Post surgery, the clinical and neuropathologic findings were all summarized (Finardi 2013). There were six males and two female with type II FCD cases. Patients one through four had epilepsy lasting one through six years and were surgically treated before age ten. Patients five through eight had epilepsy lasting 16-27 years and were treated surgically as an adult. Every patient had different results post surgery, some including an increase in cortical thickness, blurry gray-white matter junction with hyper intensity, and some had infantile asymmetric spasms at onset, and recurrent focal seizures with tonic arm posture. In all FCD patients, seizures occurred frequently when falling asleep, during sleep and on waking up. Video-EEG scalp recordings revealed interictal and ictal (seizure)

activity always related to the anatomical location of the dysplasia as indicated by MRI. Stereo-EEG recordings invariably revealed continuous interictal lesion. Cognitive function was impaired in all patients except one. All patients were followed for a long period of time after surgery. Surgical outcomes were clearly better in patients operated on in childhood than as adults (Finardi 2013). This case study proves that the sooner patients suffering from any seizures disorders, including cortical dysplasia, are operated on, the better the results are long term.

Cortical dysplasia can be diagnosed as young as infancy. Prenatal and perinatal events are suspected to be important in the pathogenesis of different types of malformations of cortical development (Krsek 2010). However, only anecdotal reports and small patients have been identified in epileptic patients with significant prenatal and perinatal brain injury (Krsek 2010). From a population of 567 children undergoing resection at the Miami Children's Hospital from March 1986 to June 2006, 200 subjects had a diagnosis of focal cortical dysplasia (FCD) and mild malformations of cortical development (mMCD). Twenty-five patients (12.5%; 9 females, 16 males) had a definite history of prenatal or perinatal risk factors according to the above-mentioned combined historical and radiologic criteria (Krsek 2010).

The study was completed on those 200 patients histologically confirmed with FCD and mMCD, including the 12.5% with definite prenatal and perinatal risk factors. The pregnancy, delivery, and postnatal histories of all subjects were obtained from obstetrical and birth records and analyzed in detail before the study was completed. Only unmistakable prenatal and perinatal risk factors were accepted. Combined historical and radiologic inclusion criteria were used to identify patients with prenatal and perinatal risk

factors (Krsek 2010). Electroclinical, imaging, neuropsychological, surgical histopathologic (examination of the brain), and seizure outcome data were all reviewed. A complete history, a neurological examination, and preoperative scalp video-EEG were evaluated in all patients.

Interictal epileptiform discharges and ictal EEG patterns were classified as regional (appearing exclusively over a single lobe or in two contiguous regions), multilobar, hemispheric, or generalized. Selected children also underwent additional diagnostic tests including positron emission tomography (PET) and single photon emission computed tomography (SPECT). A PET is a functional imaging technique that produces a three-dimensional image of functional processes in the body. A SPECT is functional imaging technique that uses gamma rays. Prenatal and perinatal results included severe prematurity, asphyxia, bleeding, hydrocephalus (when fluid accumulates in the brain), and stroke occurred in 12.5% of children with FCD/mMCD. Their epilepsy was characterized by early seizure onset, high seizure frequency, and absence of seizure control. Patients with significant prenatal and perinatal risk factors had more abnormal neurologic findings lower intelligence quotient (IQ) scores, and slower background EEG activity than FCD/mMCD subjects without prenatal or perinatal brain injury. MRI evidence of cortical malformations was identified in 74% of patients (Krsek 2010).

All the patients underwent a resective surgery for intractable epilepsy at the Department of Neurological Surgery, Miami Children's Hospital, Miami, Florida. Most patients underwent large multilobar resections of hemispherectomies; 54% were seizure free two years after surgery. Histologically "milder" forms of cortical malformations were observed most commonly in the series (Krsek 2010).

Twelve clinical parameters were compared between two groups of FCD/mMCD patients with and without prenatal and perinatal insults. Investigated parameters consisted of three interval variables and nine categorical variables. Categorical parameters were analyzed with contingency table analysis. Statistical comparisons between the groups were evaluated with Pearson chi-square statistic, Spearman R. Statistic, and also Fisher's exact test (Krsek 2010), which are all different examinations to test the correlation coefficients, the differences between the frequency and theoretical distribution, and contingencies of the seizure activity post surgery.

Only nine subjects had normal neurological findings. Neuropsychological data was available for 18 patients. More than half evidenced severe mental retardation (intellectually disabled). Mild intellectual impairment was found in six out of eighteen patients and borderline intelligence in one patient. Only one subject had normal intelligence. Several specific developmental disorders such as behavioral disorders, pervasive developmental disorders, or developmental language disorders were reported in some cases (Krsek 2010).

Eight cases had unilobar resections, seven had multilobar resections, and ten had a hemispherectomy. Three children required reoperation. Seizure outcome data two years from the last surgery was available for twenty-two subjects. Seizure freedom was achieved in twelve subjects, which was 54%. Engel class outcome was encountered in five cases (Krsek 2010), which is defined as a scale used to classify outcome after surgical treatment for seizure patients according to the Massachusetts General Hospital (*Engel Epilepsy Surgical Outcome*). As the data entails, each surgical outcome is different for all patients,

especially depending on the type of surgery (hemispherectomy or multilobar) and the severity of their seizure disorder.

Throughout this study there was also a comparison between FCD/mMCD patients with and without prenatal and perinatal brain injury. The hypothesis declared that there was no difference between the two groups at a highly significant level for parameters incidence of abnormal neurological finding, Neuropsychological ranking, incidence of slow background EEG activity, and extent of resection, that is, subjects with prenatal and perinatal risk factors had more abnormal neurological findings, lower IQ scores, higher incidence of slow background EEG activity, and greater likelihood of a larger resection. There was no statistical significance for other parameters (Krsek 2010).

Cerebral hemispherectomy has been available as a treatment for severe, intractable unilateral epilepsy in children since at least 1945 (Pulsifer 2004). Hemispherectomy significantly reduces or even completely arrests seizures and improves cognition and behavior. Partial or complete surgical removal and complete functional disconnection of one cerebral hemisphere leave the frontal and occipital poles in a place but disconnect from the rest of the brain by separation from the corpus callosum and upper brainstem (Bode, Firestine, Mathern, Dobkin 2005).

Hemispherectomy surgeries on patients with cortical dysplasia and other seizure disorders can be completed at a very young age. For these patients, the hemispherectomy results in not only seizure control but also relative improvement of language and other cognitive functions. Hemispherectomy surgeries have been reported to have 52-75% seizure-free outcome (Pinto, Lohani, Bergin, Bourgeois, Black, Sanjay, Madsen, Takeoka & Poduri 2014). While many studies have shown improved control of seizures and general

improvement, or at least no significant decline, in overall intellectual performance after a hemispherectomy, the major studies have rarely gone beyond measurement of overall intelligence (Pulsifer 2004). Patients post-hemispherectomy surgery have reported a general trend toward more functional independence and sometimes marked reduction in disruptive behavior. However, such improvement has been hard to quantify, because the studies have generally been based on anecdotal reports or individual clinical observations rather than standardized measure or structured interviews. At John's Hopkins Hospital, hemispherectomy has been performed for children with severe intractable epilepsy since 1968. Earlier reports described the clinical procedure and documented the neurological outcomes of some of these children (Pulsifer 2004).

After the hemispherectomy, cognitive assessments were completed. Throughout those assessments, significant differences were evident by the side of the hemispherectomy. Right-hemispherectomy patients scored higher in both receptive and expressive language than left-hemispherectomy patients. Patients that still suffered from seizures post-surgery were associated with lower IQ at follow up. This suggests that extremely low cognitive functioning is an indicator of generalized brain dysfunction with a reduced likelihood of seizure elimination, but is not necessarily an indicator of poorer cognitive outcome for hemispherectomy. The particular problems of children with cortical dysplasia have been increasingly recognized and could reflect organic impairment of the remaining hemisphere as well as cumulative developmental delay from the very-early-onset severe seizure disorder. Despite the less favorable outcome compared to others with different etiologies that used hemispherectomy as a corrective surgery, children with cortical dysplasia and their families expressed comparable satisfaction with the surgery

and rated their relative quality of life after surgery equally as high as did those with other etiologies (Pulsifer 2004).

Hemispherectomy surgeries can be life saving for people suffering from disorders that cause epilepsy. It also means patients to have to start over learning everything, down to how to eat and how to walk. Residual motor control can take biggest hit post-surgery. The degree of residual motor control differed for upper and lower extremities, with hand function being most severely impaired (Bode 2005).

Post-hemispherectomy motor outcomes also differed as a function of etiology. Those included disorders such as cortical dysplasia, perinatal infarct, and Rasmussen's encephalitis, all of which are neurological disorders. All children that received the hemispherectomy showed activations in the sensorimotor network ipsilateral (same side of the body) to the affected side of the brain (Bode 2005). Hemispherectomy does not seem to significantly increase the degree of motor impairments present pre-surgically, although direct comparisons of pre- and post-operative status have not yet been performed. Despite the removal of the entire sensorimotor cortex in the epileptic hemisphere, after the hemispherectomy, patients often demonstrate residual of the proximal muscles of the contra lateral upper and lower limbs and, more rarely, in the distal (hands, feet, etc) muscles (Bode 2005). With lots of therapies, patients post-hemispherectomy surgery is likely to gain back all motor control and the ability to live a seizure-free life.

### **Researcher Stance**

Given a personal experience with someone suffering with cortical dysplasia, I decided to complete a case study on a student named Connor Smith (pseudonym), who is

suffering from cortical dysplasia. The hope is to better educate others on what life is like for someone suffering from this rare disorder.

During the implementation of this case study, I observed Connor in his classroom and home setting. Throughout the observations, I looked at the type of behaviors Connor displayed due to his disability. Some could be disruptive and others involved improper uses of language or phrases to express himself. All have lead him to have an informal functional behavior assessment (FBA). During the time I was in his classroom, the FBA was revamped in order to better properly examine his behaviors. Interviews with his teacher and parents were also conducted for more information.

Upon entering the special education field, I noticed that there is a tendency for people to categorize disabilities all into one, when in reality, every disability is different. A case like Connor's it allows people to see just how individualized special education needs to be.

### **Design and Data Collection**

This case study was set up primarily on observations. I was able to observe Connor at school in his sixth grade special education class as well as his home environment. Interviews were completed in order to get other perspectives on Connor at home and at school. Data from the FBA was collected in order to see if Connor's behaviors due to his disorder could be controlled and/or decreased. Because I only had 50 hours in Connor's classroom, I was not able to see much of the implementation of the FBA. The only data available for that deals with the trial period, which was successful. In turn the plan was put into place for Connor to use for the rest of the school year. Interviews of Connor's mother and his current teacher were given in order to get different perspectives on Connor.



## **Description of Participants**

Connor Smith was the main participant in this study. Connor suffers from cortical dysplasia, which resulted in him receiving the hemispherectomy back in the summer of 2010. Due to his disorder, he has hemiparesis, mild cerebral palsy on the left side of his body. Unfortunately, the hemispherectomy was not completely successful, because about a year after his surgery, Connor started to have seizures again. As a result of his surgery, Connor displays behaviors that are almost “tic” like. Those include screaming, clapping, tapping, hitting etc. His behaviors also include use of negative comments when he is trying to express himself. For example, he told the behavior specialist to “pipe down woman,” when what he really was trying to say was “be quiet.” A FBA was put into place in order to help manage these behaviors.

His mother, Jane Smith (pseudonym) and his teacher Lori Hope (pseudonym) completed interviews with me. Jane was able to give me more insight on Connor before and after his surgery and what he is like in his home environment. Lori was able to give me insight on Connor’s academics and how he is in school. John Morgan Middle School in the Fairview School District (pseudonym) allowed me to complete my 50 hours of practicum work in Lori Hope’s sixth grade special education classroom. This allowed me to observe Connor in his classroom setting and get a stronger idea how he outside of home.

## **Evaluation/FBA**

During my time in Connor’s classroom, I was able to collaborate with the behavior specialist and Connor’s teacher in order to come up with a new FBA. The plan was focusing on all of Connor’s behaviors, “tics” and negative comments, all in one.

It became confusing for Connor's para-educator to decipher the difference and for Connor to understand what the expectations were for him. In order to make Connor's plan more successful, the FBA was revamped to focus on both Connor's "tic" like behaviors and the negative comments separately. This way both were measured. The FBA had a section that focused on Connor's ability to work quietly, meaning no noises were made throughout a lesson, and another section that focused on his respectful language. He is allowed two warnings for each behavior before he it becomes marked.

Since I was only in Connor's classroom for a short period of time, I was only able to observe four days worth of data during the trial period of this particular FBA. When Connor displayed inappropriate language, we reminded him of what language he could use. So instead of telling him the phrase he used was wrong, we would simply correct it with the proper way of expressing what he needs or wants.

During that time, Connor had a high success rate. He was about to receive a score of 96% on day one, 90% on day two and three, and 88% on day four. This created a mean score of 91%. This proved that this FBA was successful. It allowed Connor's para-educator to focus on Connor's behaviors separately, which made it easier for her to document. It also allowed Connor to have a better understanding of what the expectations were, all of which allow him to be more successful in the classroom. The hope is that this informal FBA will be used for the rest of the school year to help Connor learn how to modify the noises he makes and how to use more appropriate language when trying to express himself.

## **Interviews**

Jane Smith (pseudonym), Connor's mother, gave deep insight on the life that Connor has lived. Right off the bat, Jane noticed something wasn't right with Connor. The second

she gave birth, on June 25, 2002, she noticed that his cry was different. It sounded more like a loud shriek than a typical newborn cry. Then, when Jane went to change his diaper, she noticed he did something strange with his hands. He moved them around in a unique fashion. As a newborn, Connor went an unusually long time without food, sometimes more than five hours. Jane said she just felt in her gut that something wasn't right. "I knew something was wrong, but I just couldn't figure out what it was," (Jane Smith, April 1, 2015). Not even 36 hours after Connor born, the doctors started to notice something wasn't right too.

Jane stated, "I remember waking up around 3:30 am, noticing that Connor was still not in my room. I was about to step into the hallway when four doctors approached me. I asked where my baby was and the doctors told me they needed to talk to me. They could not tell me what was wrong, just that something was not right. They just told me that there were medical things going on and he would need more testing. So I decided then that I wanted to transfer Connor from Rochester General Hospital to Strong Memorial Hospital. He had to be taken in an ambulance but I was not allowed to ride with him. My husband, John Smith (pseudonym), had gone home to get stuff ready for Connor and I to arrive at the house the following morning. Once I got a hold of him, he came back to Rochester General and we drove behind the ambulance. Connor was then brought to the NICU. From that moment on, our lives changed forever," (Personal communication, April 1, 2015).

After multiple tests and ruling out of all kinds of disorders, the doctors finally came to the conclusion that Connor had cortical dysplasia. Once the doctors were able to put a name to Connor's disorder, it was easier to determine what kind of trials Connor was going

to face the rest of his life. One of the side effects of cortical dysplasia is hemiparesis, which is cerebral palsy on one half of the body. For Connor, since the right side of his brain is what suffers from cortical dysplasia, the left side of his body is what is affected with hemiparesis. This created him to start occupational and physical therapy at two months old.

Another huge side effect to cortical dysplasia is seizures, which meant that the unusual activity Jane suffered during pregnancy was in reality Connor having seizures inside the womb. This part of cortical dysplasia is what affects Connor the most. When the doctors were in between trying to figure out what kind of medication to put Connor on to keep the seizures at a minimum, Connor had a really bad episode of seizures. At one point, he was seizing every three minutes. It wasn't until the proper medication was identified that the seizures started to get under control. From that point on, Connor was seizure-free, at least visibly, from 18 months to age four.

When Connor was four, a family member was put in charge of Connor and his older brother, Chris (pseudonym), while Jane and John went to Florida for a vacation. Connor ended up having a grand mal seizure. From that point on Connor faced a life that involved multiple and frequent seizures. As Connor got older, the seizures become more frequent and his postictal time became very long. It sometimes took up to two days for Connor to recover. He would lose the ability to walk his ability to talk decreased. Jane described it as sounding like marbles were in his mouth when he tried to talk. His words became all jumbled and his mouth was unable to put them together. Along with the frequent seizures, Connor had a difficult time with academics. He had an extremely hard time being able to

read. His cognitive skills were very poor; overall school was very difficult for him. He had minimal ability to retain material.

When Connor was eight, the Smith family started to reevaluate his quality of Connor's life. School was becoming more and more difficult, the seizures were happening more frequently - multiple times a week - and even Connor started to grow tired of this life he was given. He started to tell his parents that he wanted the "bad things" taken out of his head. From that moment on, Jane knew she had to do something.

The first step involved Connor having long-term EEG testing done. He was brought to Strong Memorial Hospital and taken off all of his medications, including Carbamazepine, which is the main seizure medication Connor had been on his whole life. The goal of these tests was to see how Connor's body reacted when he had no medication in him. The doctors were aiming for Connor to have about five seizures, in order to look at the EEG results and the brain activity he displayed during them. That would allow the doctors to determine if Connor qualified for the life changing hemispherectomy. About nine days into his medication-free hospitalization, a seizure finally hit.

This was the most intense, long-lasting seizure Connor had ever experienced. The doctors could not get Connor's convulsions under control. They gave him Diastat (anti-convulsion medication) to stop the convulsions; after five minutes, they did not stop. They then gave him Valium (anti-convulsion medication); after five minutes of that medication being into Connor's system, the convulsions still did not stop. They then gave him Dilantan (anti-seizure medication); about five minutes after that, the convulsions started to decrease.

It was about a half hour until the convulsions completely stopped. The overall seizure lasted about two hours. The doctors were amazed, telling Jane they have never seen a seizure like that in all the years they have practiced medicine. It was never more obvious that Connor not only qualified for this surgery, but that he needed it to save his life. It was then that Jane started to research hospitals around the world to figure out which place would give Connor the best results for his hemispherectomy.

Jane researched options all over the United States, looking at different hospitals and surgeons to see which would be the best choice for her son. She came up with a questionnaire of criteria that she used to help her narrow down her design. It involved how the surgery is handled, postsurgical “laying down” of the patient, postsurgical rehab, infection rate per surgeon and how many hemispherectomies the surgeon had performed. In the end, Jane decided to go with a surgeon at Strong Memorial Hospital who had given thirteen hemispherectomy surgeries in his career.

Connor had a functional right hemispherectomy surgery on July 19, 2010. He was eight years old.

That day, Connor spent twelve hours in the operating room of Strong Memorial Hospital. The surgery was only supposed to take eight hours, but due to hemorrhaging that required three transfusions, the surgery ended up taking a lot longer than planned. Since Connor had a functional hemispherectomy, the parts of his right side of his brain that were not malformed were left inside the head. Those were the frontal and occipital lobes. The corpus callosum was disconnected because Connor’s seizure activity misfired so rapidly, that if the connection was left, the activity was most likely going to transfer to the left side of the brain. This would, in turn, cause a seizure.

When the surgeon came out of surgery, there was a huge crowd in the waiting area anxiously awaiting good news. Thankfully, everything ran smoothly, despite his hemorrhaging and need for transfusions. It didn't take long before people lined up to go see Connor, who spent twelve days at Strong Memorial Hospital before he was transferred to Saint Mary's Brain Injury and Rehabilitation Center. Here Connor spent the rest of the summer learning how to talk, walk, and strengthen the left side of his body all over again. It was the most difficult trial that Connor and his family had to face. He had three to five hours a day of pure therapy sessions. Once he was able to come home, Connor still faced therapy sessions three hours a day.

Due to his high therapy needs, Connor did not return to school full time until June for the 2010-2011 school year. During that time, a tutor hired by the Fairview School District came to Connor's home and taught him mathematics and reading.

His ability to learn improved dramatically. Connor was able to retain what was being taught. He was able to start reading, which was a noticeable improvement, according to Jane and the tutor. Another factor as to why Connor did not go back to school until June was because he suffered from posttraumatic stress symptoms, which is a side effect of traumatic brain injuries. He would get very anxious in surroundings with crowds of people because his awareness increased post-surgery, creating anxiety that was not there before. This caused Connor to suffer from anxiety not only in unfamiliar settings, but in settings that he has been around for years. It has been a huge adjustment for Connor to get used to large crowds again.

There are many other issues that develop as a results of hemispherectomies. Before surgery, Connor had very typical behavior for his age group, despite the learning disability,

hemiparesis and seizure disorders. Post-surgery, the first thing the Connor's doctors and family noticed was that his left hand was flat, which was something that never happened pre-surgery. Connor was unable to open his hand completely before so this was a great sign. It caused doctors to be very hopeful for the future, but only time would tell. One of the most noticeable side effects to Connor's surgery was his behavior. Post-surgery, he developed the tic-like behaviors, described earlier. These behaviors tend to come about when he is anxious or over-stimulated. Connor also has a tendency to use what is referred to as negative comments or inappropriate phrases when trying to express himself, mostly due to that fact that he is unable to think quickly enough before saying what comes to mind, no matter how outlandish the language may be. As noted, because of this, he has an informal functional behavior assessment in his sixth grade classroom.

At first, everything seemed to be improving, especially the seizures. From what the doctors could tell, there had not been any seizure activity post-surgery. That quickly changed, as Connor started seizing again - except these seizures looked different. Connor kept telling people he felt "dizzy," which was unusual. Jane knew something was not right. Connor came home from rehab in August 2010 and ended up back in the hospital in September 2010 because of the complaints. About ten months post-surgery, things started to get worse. Connor started having seizures that became noticeable to Jane. Doctors did tests to see how this was possible, considering the entire right hemisphere of his brain, where the malformation was, had been removed.

Sadly, the doctors determined was that there is a small mass in the frontal lobe of Connor's brain that was not removed due to the amounts of hemorrhaging. The surgeon simply did not see it. This small mass in Connor's brain is still causing seizures, which in



turn, have made it so the surgery was not as successful as Jane and the surgeons had hoped. Jane stated, "I was happy with the decrease of seizures in occurrence and intensity. Overall the surgery improved his ability to learn, which he did not have before. Some of the side effects make it very frustrating, and it also causes you to lose part of that person. There are still parts of Connor there. The animated Connor is gone, which has been the most difficult part" (Personal Communication, April 3, 2015).

With the complications that have arisen over the past few years, there was talk of Connor having surgery again to remove that small mass in the frontal lobe of his brain. It was eventually scheduled for August 12, 2013. On August 6, 2013, Connor was watching a neighborhood football game when a player accidentally ran into him, causing him to fall down patio steps, which severed his left ear. Due to this accident, Connor was bleeding internally, which meant the doctors had to cancel his surgery. Right now the thought of Connor having surgery again is up in the air. Jane stated, "It will be based on whether the seizures gets worse and whether or not he shows signs of the activity going left-sided, although the number of seizures has increased within the past month or so," (Personal Communication, April 2, 2015). Since he started going through puberty, the brain is maturing which can cause the activity to change. Once that process is completed, it will be easier for doctors to tell if the surgery is necessary or not. Jane is hopeful that Connor will have a bright future, despite the challenges he will forever face.

During my practicum, I was able to work closely with Connor's teacher, Lori Hope (pseudonym). Lori described working with a student like Connor as "awesome." He's an amazing young boy that has overcome so many challenges at such a young age. His love for his family and friends shines through him, along with his desire for independence and

success in school. Despite how wonderful Connor is to have in her sixth grade special education classroom, the inconsistency of his behaviors can make it very challenging to have him in the classroom. Due to Connor's disorder and surgery, as mentioned before, he tends to make random noises and negative comments, randomly throughout the school day. It is very difficult for Lori to manage these behaviors in a way that will create success for Connor. It is difficult being able to know if these behaviors are purposeful or if Connor cannot control them. For Lori, this has been an ongoing process to figure out throughout the school year.

Fortunately, the new FBA that has been in place has been successful at helping Connor understand what the behavior expectations are and how to appropriately use phrases to express himself. The educators in the room are better able to document the noises and negative comments that Connor makes. This increases Connor's ability to be aware of his own behaviors, as well as being able to inform Jane how he is doing in school too.

As apart of the FBA, the educators are able to reinforce appropriate phrases to show Connor how he should express himself. Instead of telling Connor what he said wasn't appropriate, the overall idea is to remind Connor of what phrases to use when he is feeling a certain way. It took a lot of trial and error before Connor's FBA was correctively gathering data and focusing on helping Connor use appropriate phrases. The recent FBA has effectively allowed Connor to be able to monitor the noises he makes and teach him appropriate phrases to use instead of the negative ones.

Working with a student like Connor has been an entirely new experience. Learning the most effective ways for dealing with his behaviors has been a day-to-day trial. What

Lori has noticed has a higher success rate is allowing Connor to get the noises out of his system before school starts. This way Connor has the ability to get it out of his system. Other times, a direct, straightforward approach is necessary. Distracting him, using humor, and switching up where and who he works with seems to be effective. Connor receives positive attention very well. The behavior plan also is effective. Not all of these approaches work all day, every day, but overall, switching up these approaches seem to help Connor be successful in the classroom.

Connor is not Lori's first student with traumatic brain injury (TBI). Last year she had one student that suffered TBI from a car accident. This was a completely different experience. This student had a wheelchair because of her accident. Despite her physical limitations, she still was capable of learning positive behavior strategies and applying them to new situations. She is also able to retain academics more effectively. Despite how different these two cases with TBI are, both students have a great attitude, and persevere when tasks are challenging, whether the challenge is physical or academic. Both experiences with these students have been rewarding and challenging.

The part that Lori has found to be the most beneficial experience to having a student with such a rare and challenging disorder was learning how valuable the parents' knowledge, insight, and experience can be. With every year that Lori's teaches, she continues to learn that this is true. However with Connor's case, the more Lori was able to learn about him, the better she was able to understand him and the things that he does. Since the insight his parents have given Lori has been so beneficial, she has been able to pass on that knowledge to other people at Connor's school in order to help them in working with him. This, in turn opens up a path for Connor to be more successful.

Connor can be a very difficult student to understand due to the side effects of his surgery and disorder. In turn, it takes people with a lot of patience and understanding to effectively work with him. Since Connor has such high needs, he does have a para to assist him throughout the day. Lori states that working with a para can be a little bit of both rewarding and challenging. When it comes to a case like Connors, working with a para has been more challenging this year, not by any fault of the para, Lori, as well as myself, feel that para's need to be trained in the area of the student's disability. Unfortunately, that is not something Connor's school offers. Because of that, educating the para falls onto the teacher.

Lori stated, "We barely have any time to talk to the para's, and having time to discuss the disabilities, Individualized Education Programs (IEPs), and behavior management plans is just impossible," (Personal Communication April 6, 2015). Despite the challenges to find time to discuss Connor with his para, Lori has found that she is ready to learn and take constructive criticism. The biggest challenge is that Connor's para's style of interacting with students is different than Lori's. Despite that, it is easy to tell that she cares a great deal for Connor. She knows him well and knows what activities he needs support with and what activities he can work on independently. Knowing this sets up Connor for success throughout his day.

Academics have always been difficult for Connor. He learns very differently than most students, meaning he needs assistance or alternative ways in order to help him learn most effectively. Connor can focus best when his desk is close to the front of the room, away from other students and near Lori's desk. He needs larger lines on his papers because his writing skills are poor. The iPad has become a major tool that Connor uses in the

classroom. He has a talking calculator, extra wait time (especially due to his slow processing skills) a page marker, Post-it notes, one-on-one support and the behavior plan. All of these modifications allow Connor to work to his best ability. He is able to come to school and enjoy himself. Even if certain subjects are challenging, he still works hard and tries his best to be successful.

### **Summary**

As Connor's cousin, I have been fortunate to get a front row seat to his life story. I'll never forget the day I went to visit him as a newborn. It was shortly after he came home from the hospital. My father was holding him when he suddenly began to seize in his arms. Connor's face turned bright red, his body began to twist and turn in different directions while my father remained still until the seizure stopped. The room fell silent instantly, everybody stopped and took a deep breath. It was then, even being 13 years old; that I realized the challenges Connor was going to face for the rest of his life.

Due to cortical dysplasia, Connor has faced many trials so far in his life. I'll never forget watching him participate in therapy, whether it was occupational therapy, speech therapy and physical therapy, at such a young age. Despite all of the challenges Connor faced, he was always positive and full of laughter. He was one of the most playful, active and full of life children around. He could always bring a smile to someone's face. One of the things I admire most about him is he never was phased by only being able to use the right side of his body to its best function. Doing things one handed became the norm for him. It was amazing to observe him learn how to play sports and other activities using primarily one side of his body. He made it look so easy.

I spent a good portion of my teenage years babysitting Connor and his older brother Chris. Due to that, I was taught what procedures to do in case Connor had a seizure while I was babysitting. One of the essentials was making sure Connor got his medication every night. Without it, he could start seizing and lose control of his body. Being exposed to someone with such a rare disability at such a young age was truly a life-changing experience. It opened up my eyes to see just the kind of trials people with disabilities face.

From that moment on, my curiosity towards special education grew. It was from my personal experience with Connor that allowed me to see where my passion lied. Connor is such an inspirational child. He has been through so much in such a short period of time but still manages to have such a positive outlook on life. It is because of him that I knew special education was the right career choice for me.

Completing this case study has expanded my knowledge on cortical dysplasia and the challenges Connor faces on a daily basis. It's given me a whole new perspective on how people with disabilities function. Everything is seen through a different pair of eyes, which in turn results in different ways of learning. As an educator, this experience has taught me just how important differentiation is. It takes a whole new meaning after observing a student like Connor. Everything has to be individualized in order for him to learn effectively, which causes me to see how essential it is to get to know my students the best I can. In turn, it will result in me learning what teaching styles and strategies work best for my students. This sets up the students for a higher success rate.

Parents of a student like Connor also play a huge role in their success. I was able to see just how beneficial the parents insight, knowledge and experience are not only for the

student, but for the teacher as well. All of which are important factors for a successful school year.

Without Connor, I would not have had the opportunity to watch an extremely brave child embark on life-changing situations with a bright smile on his face. It is because of him that I am better able to understand the trials students with disabilities face, and for that I thank him. People like Connor remind us just how amazing life can be, and if anything has come out of this case study, it's knowing that no matter how we look at life, we all have a reason to smile.

### **Next Steps/Follow Up**

Connor is finishing up his sixth grade year at John Morgan Middle School in the Fairview School District. He will continue school through the summer too. Unfortunately, his seizures have recently become more frequent. Surgery is still up in the air and may not be determined until a later time. Connor's family and doctors are keeping a close eye on him as he continues to grow. Academics will continue to be struggle for Connor, but with the necessary modifications and individualized plans, he is able to achieve success.

There is no way to know what the future holds for Connor, but all are hopeful that with or without the surgery, he will continue to live a fun-filled life with minimal seizures. Despite recent challenges, Connor continues to have a bright smile and positive attitude. No matter what trials he faces, that side of him will always remain to be what makes Connor such an inspiration.

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